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## CASE REPORT

# Juvenile ossifying fibroma of the maxilla

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### KEYWORDS

Juvenile ossifying fibroma;  
Cemento-ossifying fibroma

### Summary

**Introduction:** Juvenile ossifying fibroma is a rare, benign fibro-osseous tumour. In the light of a clinical case, the authors review the diagnosis, treatment and histological classification of these tumours.

**Case report:** A seven-year-old child presented with a naso-orbito-ethmoidal trabecular juvenile ossifying fibroma. Complete surgical resection via a transfacial approach was performed after a preoperative work-up comprising head and neck CT and MRI.

**Discussion:** A review of the literature reveals that treatment of this aggressive tumour must comprise complete surgical resection via an incision determined by local tumour extension.

**Conclusion:** Patients with juvenile ossifying fibroma tumour require long-term follow-up due to the high recurrence rate.

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## Introduction

Juvenile ossifying fibroma is a rare histological entity belonging to the group of fibro-osseous tumours [1]. This benign tumour of the facial bones classically arises in the nasal sinuses and can present intracranial and orbital extensions [2]. Its aggressive and osteolytic nature due to intense osteoblastic activity [3] can simulate a malignant tumour. Juvenile ossifying fibroma is often difficult to distinguish from other fibro-osseous lesions on clinical examination and complementary investigations. The authors report a case

of trabecular juvenile ossifying fibroma in a seven-year-old girl.

## Case report

A seven-year-old girl, A. B., with no personal or family history, consulted for painful, complete right nasal obstruction present for two months associated with right exophthalmos and diplopia. Physical examination revealed deformity of the orbital region and confirmed nasal obstruction by a mass lined by normal mucosa. Ophthalmological examination revealed moderate diplopia with no signs of oculomotor paralysis and no loss of visual acuity or visual field abnormalities. Computed tomography (CT) of the facial bones (Fig. 1) demonstrated a relatively well circumscribed cal-

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**Figure 1** Computed tomography, tissue window setting. A. Coronal section. B. Sagittal section.

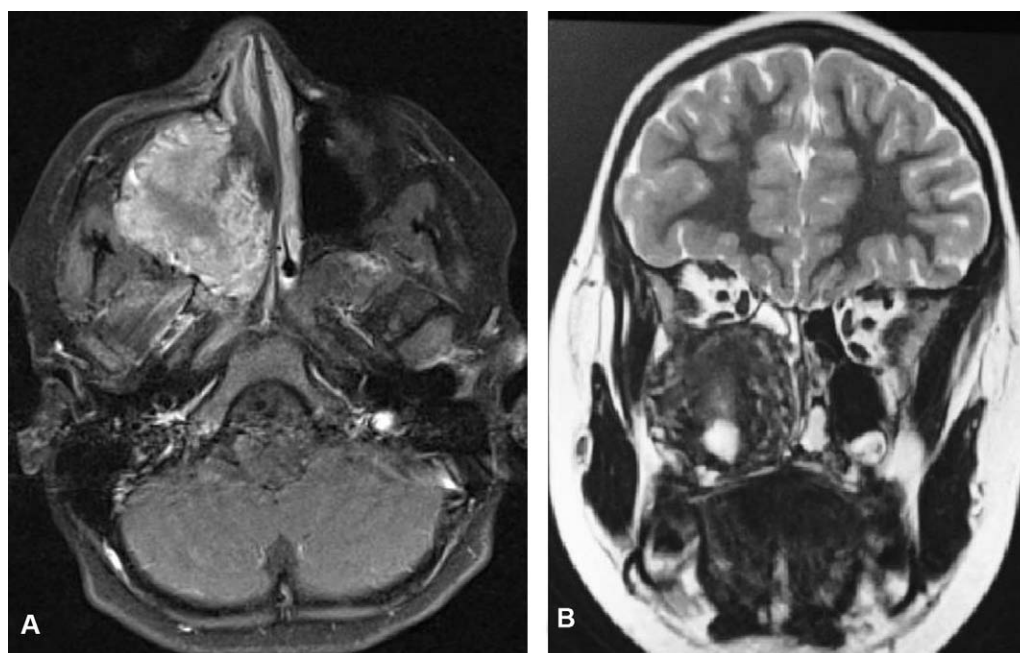
cified lesion, displacing the adjacent structures of the right maxillary sinus and nasal septum, inducing osteolysis of the orbital floor and right ethmoid bone, with no involvement of the roof of the ethmoid. This heterogeneous tumour measuring  $3.5 \times 4 \times 4.2$  cm was well circumscribed on MRI (Fig. 2), which showed intraorbital invasion in contact with the optic nerve and osteolysis of the posterior wall of the maxillary sinus and the pterygoid process.

A biopsy was performed under general anaesthesia through an endoscopic approach and showed a trabecular juvenile ossifying fibroma. The tumour was resected via a transfacial approach (Fig. 3) justified by the invasion of the orbital floor and pterygoid process. This procedure allowed *en bloc* resection of the tumour with preservation of the orbital periosteum and reconstruction of the orbital floor with a Silastic® implant. The patient experienced transient

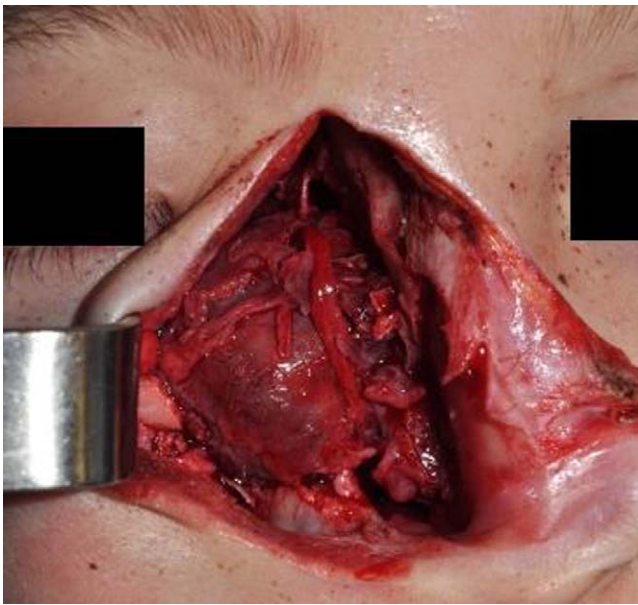
postoperative diplopia and no recurrence was detected on follow-up imaging performed nine months after the operation (Fig. 4).

## Discussion

The terminology of fibro-osseous tumours has been confusing for a long time. Cemento-ossifying fibroma and juvenile or adult ossifying fibroma were considered to be two distinct histological entities, as the first was considered to be an odontogenic tumour, while the second was not. Although this distinction was proposed in the WHO classification of 1992 [1], several authors [4,5] have considered this distinction to be arbitrary and useless, and have suggested grouping these tumours under the term "ossifying fibroma" due to



**Figure 2** MRI. A. Gadolinium-enhanced T1-weighted sequence, axial section. B. T2-weighted sequence, coronal section.



**Figure 3** Operative view of the right paralateronasal approach.

their histological similarities. The terms "psammomatous fibroma", "sclerosing fibroma", "localized hypertrophic or fibrous osteitis", "localized fibrous osteodystrophy", "osseous keloid" and "fibrous osteoma" are also used in the literature to refer to this same tumour. The term "cemento-ossifying fibroma" is still frequently used in recent articles, while the term "ossifying fibroma" appears to be reserved to sites other than the jaws. However, three forms of ossifying fibromas have now been distinguished [3]: classical ossifying fibroma, psammomatoid juvenile ossifying fibroma and trabecular juvenile ossifying fibroma. The term "cemento-ossifying", which referred to the presence of basophilic mineralized structures resembling the cementicles observed adjacent to the roots of teeth, is therefore tending to be abandoned. The dental origin of these tumours was excluded when these structures were demonstrated in fibromas of long bones.

Histologically, these lesions are always benign, composed of highly vascular and fibroblast-rich connective tissue, which produces a calcified substance that often cannot be clearly attributed to either cement or bone. Clumps of osteoblasts are also present. The differential diagnosis of ossifying fibroma with fibrous dysplasia or extracranial psammomatous meningioma can therefore be difficult.

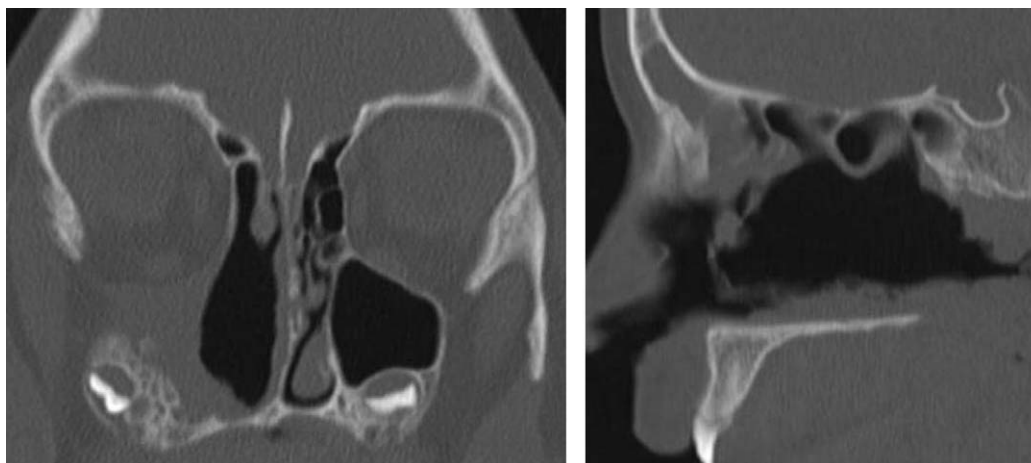
Juvenile ossifying fibroma affects children under the age of 15 years in 80% of cases [3] and arises in the orbit or nasal sinuses in 90% of cases [6], in contrast with classical ossifying fibroma which generally arises in the mandible.

The incidence of juvenile ossifying fibroma is unknown. A review of the literature revealed 17 cases reported between 2003 and 2010 with a sex-ratio of five females for one male in adults, while a male predominance is observed in the juvenile form. These lesions can be slowly evolving and asymptomatic, but appear to be more aggressive in young subjects [3]. Oukabli [7] reported the case of a 36-year-old woman with a two-year history of symptoms, while the case reported here had experienced symptoms for only two months. Pace [6] calculated a tumour doubling time of 3.5 months in children.

Radiologically, juvenile ossifying fibroma presents as a clearly circumscribed, concentrically expanding, solitary mass with bone density. It is the circumscribed nature of ossifying fibroma which distinguishes it from fibrous dysplasia. The fibrous centre of the tumour is less dense on CT with little or no contrast enhancement. On MRI, as in the case reported here, this lesion has a heterogeneous high-intensity signal on T1-weighted sequences and a low-intensity signal on T2-weighted sequences. However, the various histological subtypes of fibro-osseous tumours cannot be distinguished radiologically.

The reference treatment consists of complete surgical resection. In contrast with fibrous dysplasia, no data concerning the use of bisphosphonates are available.

An endoscopic approach could have been used for a smaller tumour, but an open, transfacial approach was preferred in this case due to the large tumour, the high recurrence rate and the difficulty of ensuring local control in the anterior part of the maxillary sinus. A degloving approach could also been used.



**Figure 4** Computed tomography nine months postoperatively. A. Coronal section. B. Sagittal section.

As postoperative recurrences of ossifying fibroma have been reported (5% for classical ossifying fibromas [8] and 25 to 58% for juvenile forms [3,9]), long-term surveillance is therefore justified.

## Conclusion

Ossifying fibroma always has a benign course but requires early surgical management due to its aggressive and compressive nature. Orbital extension, as in the present case, and intracranial extension remain exceptional. The recurrence rate justifies long-term clinical and radiological surveillance.

## Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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